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A Retrospective Study In Children With Pulmonary Arterial Hypertension: A Single Center Experience

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Objectives: To determine the epidemiology, quality of life and management and effectiveness of the treatment of pediatric pulmonary arterial hypertension (PAH).

Methods: Patients who had diagnosis of PAH and treated at Department of Pediatric Cardiology, Gazi University Faculty of Medicine from February 2006 until October 2015, were evaluated retrospectively.

Results: Forty-one patients were included with a mean age of 83 ± 68 months (21 Female, 20 Male).

85.4% of patients had PAH associated with congenital heart disease and the ventricular septal defect was the most frequently defect. Four patients had primary PAH (9.8%), 1 patient had PAH caused by chronic lung disease and 1 patient had PAH caused by obstructive sleep apnea (2.4%). The most common clinical symptom reported was fatigue (65%). Telecardiography, electrocardiography (ECG) and echocardiography (ECHO) were performed in all patients. 6-minute walk test (6 MWT) were performed in twenty-five patients (61%). Cardiac catheterization was performed in 40 patients (97.6%). Only one patient couldn't be performed catheterization due to her bad clinical condition.

Sixty percent of the patients, who had catheterization, were performed vasoreactivity test and 39% of them had positive response. There is no significantly difference between measured mPAB values in ECHO and catheterization (60 ± 53.5 mmHg vs. 65 ± 20 mmHg respectively). While the most frequently prescribed monotherapy was bosentan 48.8%, for the combined therapy it was bosentan plus iloprost with 22%. Eleven patients (26.8%) received monotherapy, 19 (46.3%) received dual therapy and 3 (14.3%) received triple therapy. During the follow up, the combined therapy was increasingly prescribed (43.9% versus 66.9%). The mean time of adding second drug was 27 months. During the follow up, 4 patients died. One- and five- year survival rates were 94% and 86%, respectively.

Conclusions: At the experienced centers, the positive results on the survival rate and life quality of patients with PAH, obtained with the current treatment options. As seen in this study, PAH is a progressive disease and the combined therapy is inevitable. Owing to this reason, the combined therapy should be considered in the early stage of the disease.